WHAT YOU NEED TO KNOW ABOUT

Wilms' Tumor

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WILMS' TUMOR

INTRODUCTION

This pamphlet is written for you, the parents of a child with Wilms' tumor, in the hope that it will contribute to your understanding of the disease and its treatment.

We believe it is important for you and your family to understand this illness so you can face it together, with courage and hope.

Much has been learned about *cancer** in general and Wilms' tumor in particular in recent years. As a result, great strides have been made in

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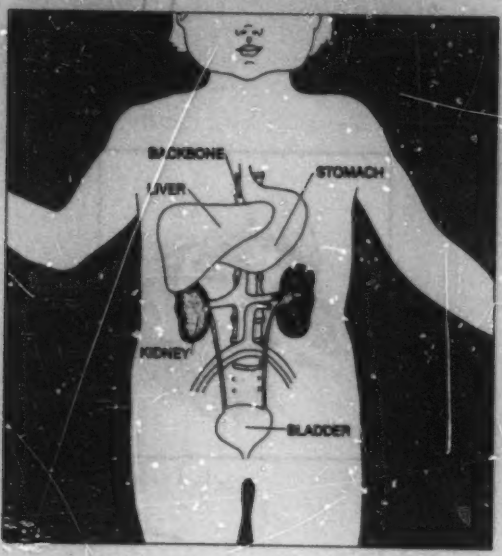
* *Words in italics are explained at the end of the pamphlet.*

treating the disease effectively. The nature of Wilms' tumor and the modern methods of treatment are described in this pamphlet. You also will find information on sources of assistance, areas of research, and definitions of medical terms.



WILMS' TUMOR

Almost without exception, cases of *kidney* cancer in children are Wilms' tumors. Scientists think this type of cancer starts in immature cells that normally become mature renal (kidney) cells.

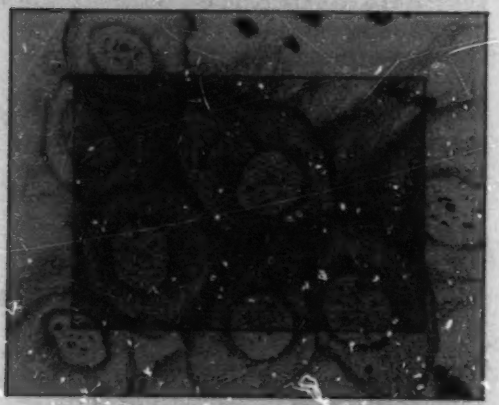
The kidneys are a matched pair of vital organs located below the liver and stomach and near the backbone on either side. Shaped like the "kidney bean," a 3-year-old child's kidney is about 2 inches long, 1 inch wide and weighs about a quarter of a pound.

The kidneys help remove wastes from the body by making urine. They do this by filtering *urea*, salt and other

substances from the blood as it flows through the kidneys.

Wilms' tumor occurs in children from infancy to age 15. It is rare in older patients and is very different from adult kidney cancer.

As with all cancers, Wilms' tumor is a disease of the cells. Cells normally grow in an orderly, controlled pattern. As cells wear out, new ones are produced. Just enough new cells grow to replace the old ones. Cells of each part of your body such as your bones, skin, and heart differ in shape and function. Each type of cell is designed



Normal cells

Cancer cells

to do a particular job in a particular organ.

When cell division is not orderly, abnormal growth takes place. Masses of tissue called *tumors* build up. Tumors may be *benign* or *malignant*.

Benign tumors remain localized and usually do not spread or threaten one's life. They can be removed completely by surgery and are not likely to recur.

Malignant tumors are cancers. They can invade and destroy nearby tissues and organs or spread to other parts of the body by way of the bloodstream or *lymphatic system*. The new growths they form are called secondary tumors or *metastases*. Even if a primary cancer is removed by surgery or *radiation therapy*, the disease sometimes recurs because it may have mestastasized before the original tumor was eliminated.

SYMPTOMS

Wilms' tumor has no unique symptom to reveal its presence. The most common sign is a lump in the belly, or a swollen *abdomen*. Blood in the urine occurs in about 25 percent of Wilms' tumor patients but in many instances is visible only when viewed through a microscope. It may be present constantly, or it may be seen only from time to time. Blood in the urine also can be a sign of disorders other than cancer. But no matter what the cause, blood in the urine should be brought to the attention of a doctor.

An occasional child will be found on examination to have high blood pressure.

Like all cancers, kidney cancer in some children can cause low grade fever, fatigue, loss of appetite, weight

loss and anemia. When you see your doctor, you should report all your child's aches and pains. They may be important clues in making a diagnosis.

You may have been alerted to the possibility of a Wilms' tumor developing in your child if another member of your family or you yourself have had the disease.

You also may have been alerted to the possibility if your child has displayed one or more of a number of so-called *developmental anomalies.*

Aniridia, or absence of the iris of the eye (the colored portion of the eye), is the most frequently encountered developmental anomaly associated with Wilms' tumor. Aniridia occurs in approximately 1 out of 173 cases of Wilms' tumor, in contrast to 1 out of 50,000 persons in the overall population. A condition called *hemihypertrophy,* or an abnormal enlargement of a part of the body, is the next most frequently encountered anomaly in Wilms' tumor patients. Abnormalities in the development of the genitals or the urinary system also appear in Wilms' tumor patients with greater than normal frequency.

The chances are that you, the parent, will have been the first person to detect the actual presence of a Wilms' tumor. You may have noticed a slight swelling in your child's abdomen, or you may have felt a hard mass in his abdomen while bathing or dressing him.

Because the risk of metastasis (spread of the cancer) increases with time, masses of undetermined origin should be treated promptly. Most Wilms' tumor patients will begin treatment very soon after their visit to the doctor, with diagnostic and

preoperative tests scheduled on a first-priority basis.

DIAGNOSIS

Diagnosis begins with a physical examination and a review of your child's medical history. Diagnostic radiology (pictures taken with *X-rays*) is always used to confirm a suspected diagnosis of Wilms' tumor. The method used most often is called an IVP, which stands for *intravenous pyelography*. A substance that can be seen in an X-ray picture is injected into a vein, flows through the kidney, and comes out in the urine. The substance "lights up" the kidney in X-ray pictures.

The doctor also may arrange for a special kind of X-ray test called *nephrotomography* (tomography of the kidney). Tomography provides a series of pictures. When the pictures are put together, they can give a three-dimensional picture of the kidney and any associated tumor. A type of tomography introduced recently is called the CAT scan, an abbreviation for computerized axial tomography scan. While your child lies still, a narrow X-ray beam directed by a computer revolves around him or her. In a matter of seconds, the machine registers thousands of bits of information that are translated into a cross-sectional picture on a viewing screen.

In addition, a "sound-picture" of your child's kidney probably will be made with *ultrasound*, a technique based on the principles of sonar.

Nephrotomography and ultrasound can tell the doctor with a very high degree of accuracy whether a

suspected mass is a *cyst* (a fluid-filled sac) or a tumor. If the first tests are inconclusive, another kind of X-ray called *selective renal arteriography* can tell the doctor whether the tumor is likely to be cancerous. Selective renal arteriography produces a picture of veins and arteries that crisscross the kidney and tumor. The veins and arteries are made visible by the introduction of a chemical similar to that used for the *IVP* into the bloodstream through a *catheter* placed in the *femoral artery*. This is not a painful procedure, but it requires a few hours of hospitalization and a mild local sedative.

If the radiological tests confirm a diagnosis of Wilms' tumor, the doctor will want other tests that will tell him whether the cancer has spread locally or to other areas of the body.

An X-ray examination called *venacavography* is sometimes given along with the IVP. It indicates whether and how far the cancer may have penetrated the vein that carries blood out of the kidney. The search for possible distant metastases may include chest and bone X-rays, a liver function test, and a series of *radioisotope bone scans*. The lungs, liver and the bones are the areas to which Wilms' tumors most often metastasize.

About 10 percent of Wilms' tumor patients have tumors in both kidneys. Usually one tumor is considerably smaller than the other, making it feasible to remove one kidney but only part of the other. The kidneys have enormous compensatory powers and, with reasonable caution, it is possible to lead a normal life with only part of one kidney intact. In the rare case that both kidneys have to be removed, a

kidney transplant or artificial kidney will be needed.

When a diagnosis of Wilms' tumor has been confirmed, it is best for your child to begin treatment in a hospital that has an expert pediatric staff and the resources to apply all forms of effective treatment right from the beginning. You may wish to request a second opinion from another physician to confirm the first diagnosis and recommendations for therapy.

TREATMENT

The discovery and development of effective treatments for each of the more than 100 types of cancer is a major goal in the national effort to bring cancer under control. Wilms' tumor is one of some 20 cancers for which treatments have been developed combining surgery, *radiation therapy* and *chemotherapy* (treatment with anticancer drugs). Overall, more than 8 out of 10 youngsters with Wilms' tumor can expect to achieve the long-term disease-free status that, for this cancer, is the equivalent of cure.

The way in which the three treatment methods will be used in

your child's treatment depends upon his medical history and general health, but above all on the stage of his disease. Radiation therapy, for example, is not used in children under the age of 2 when they have localized tumors.

The surgical procedure used in the treatment of kidney cancers is called a *radical nephrectomy*. The word "nephrectomy" means removal of the kidney. "Radical" means that large portions of the tissue surrounding the kidney and the neighboring lymph nodes are also removed.

In recent years, advances in surgical techniques and medical care have made extensive surgery possible for children and infants previously considered too young for this treatment. Today surgeons have the help of highly competent teams of anesthesiologists, nurses, therapists, and other professionals to support patients during surgery and throughout their postoperative period.

When radiation therapy is used after nephrectomy, its purpose is to guard against recurrence of the tumor where the kidney was removed. The

basic principle of radiation therapy is to restrict the beam of radiation to the area at risk for tumor growth, at doses that will destroy cancerous cells without causing undue damage to surrounding normal tissue. Radiation therapy uses X-rays, gamma-rays from cobalt-60, or other kinds of ionizing radiations.

Occasionally, when a Wilms' tumor is too large to be immediately operable, radiation therapy or chemotherapy will be used to shrink the tumor. Preoperative radiation usually involves a waiting period of up to 4 weeks between preliminary treatment and surgery.

Chemotherapy is used to treat virtually all cases of Wilms' tumor. Anticancer drugs kill cancer cells. Because the drugs act on normal cells as well as cancerous ones, the doctor must maintain a delicate balance of enough drugs to kill cancer cells without destroying too many healthy cells. The length and frequency of chemotherapy depend upon the stage of your child's disease, how long it takes him to respond to these drugs, and how well he tolerates the side effects of the drugs.

No element of the treatment regime for Wilms' tumor is entirely free of side effects. Surgery, radiotherapy and chemotherapy all can contribute in varying degrees to suppression of the body's natural immune defenses and blood-coagulating capabilities, thus subjecting the child to short periods of risk to infection and bleeding. Radiotherapy can cause a temporary inflammation of the remaining kidney. Complications associated specifically with chemotherapy may include alopecia (hair loss), impairment of nerve function, nausea

and, possibly, liver and heart dysfunction. Some patients will experience only one or two of the conditions; others will not be affected at all. These side effects are almost always short-lived, manageable and reversible.

Radiation therapy can affect growth and development of the spinal column in some patients. Radiation also slightly increases the patient's chances of developing new cancers years in the future. Younger patients are more susceptible to the effects of radiation on the spine, because bone growth is incomplete. One of the major recent advances in treatment research has been the discovery that chemotherapy can be substituted for radiation therapy, at least in babies and children under 2 when the Wilms' tumor is localized.

AFTER TREATMENT

The doctor will discharge your child from the hospital as soon as possible. Most often he will be well enough to lead a normal life at home and school.

It is important to discuss with the doctor just how active the child can be so that he will not be overly limited in his play or attendance at school. To overprotect a child in this situation may unnecessarily keep him from many of the activities that he enjoys the most. Also, to alter disciplinary measures or to overindulge the child with toys and other gifts may lead to a situation that is unrealistic from the child's point of view. Children need certain limits and boundaries and are usually happiest when treated in a normal and consistent manner.

On the other hand, the doctor may feel that the child needs special care and consideration. If these are necessary, the overall program may be briefly explained to him so that he does not view these limitations as punishment. It should be made clear to the child that if he becomes ill and has to return to the hospital, it is not because he did anything "bad." The hospital and the child's doctor should always be pictured as a source of help.

If there are other children in the family, they will want to know what is wrong with their brother or sister. They will sense from the actions of their parents that something serious has happened. It is important to answer their questions honestly. While younger children may be satisfied with simple explanations, older children will want more detailed information. It is important to indicate that Wilms' tumor is serious and not "just a cold." Children can be assured, however, that the disease is not contagious and that they need not worry about their own health. When children understand the illness, they are less likely to resent the special treatment which is invariably given to

the ill child. In addition, older children welcome the opportunity to be taken into the family confidence and will often respond in helpful ways. In, fact, the opportunity to participate in some manner in the care of their brother or sister gives them a sense of belonging which tends to promote family solidarity.

The social service department of the hospital can advise parents about local organizations that offer help for cancer patients and their families. Many organizations provide services that may include financial aid, transportation to and from the hospital for medical care, person-to-person assistance through group meetings, as well as other services.

Families who have been through it may be helpful in dealing with Wilms' tumor. They may be reached by contacting the Candlelighters, an organization of parents of young cancer patients, at 123 C Street, S.E., Washington, D.C. 20003.

In addition, the Cancer Information Service is a toll-free telephone inquiry system which supplies information about cancer and cancer-related resources to cancer patients, their families, and health professionals. Cancer Information Service offices are based in major cancer programs around the country.

For answers to questions you may wish to ask about cancer or resources in your area, you can call the telephone number for your state listed below.

ALABAMA: 1-800-292-6201
ALASKA: 1-800-638-6070
CALIFORNIA:
From Area Codes (213), (714), & (805) 1-800-252-9066
COLORADO: 1-800-332-1850

CONNECTICUT: 1-800-922-0824
DELAWARE: 1-800-523-3586
DISTRICT OF COLUMBIA:
636-5700
FLORIDA: 1-800-432-5953
GEORGIA: 1-800-327-7332
HAWAII:
Oahu: 524-1234
Neighbor Islands: Call Collect
ILLINOIS: 800-972-0586
KENTUCKY: 800-432-9321
MAINE: 1-800-225-7034
MARYLAND 1-800-492-1444
MASSACHUSETTS: 1-800-952-7420
MICHIGAN: 1-800-482-4959
MINNESOTA: 1-800-582-5262
NEW HAMPSHIRE: 1-800-225-7034
NEW JERSEY: (Northern)
800-223-1000
NEW JERSEY: (Southern)
800-523-3586
NEW YORK STATE: 1-800-462-7255
NEW YORK CITY: (212) 794-7982
NORTH CAROLINA: 1-800-672-0943
NORTH DAKOTA: 1-800-328-5188
OHIO: 1-800-282-6522
PENNSYLVANIA: 1-800-822-3963
SOUTH DAKOTA: 1-800-328-5188
TEXAS: 1-800-392-2040
VERMONT: 1-800-225-7034
WASHINGTON: 1-800-552-7212
WISCONSIN: 1-800-362-8038

ALL OTHER AREAS: 800-638-6694

RESEARCH

The National Cancer Institute, a bureau in the U.S. Department of Health, and Human Services, is the Federal Government's principal agency for cancer research. The Institute supports research in many of the nation's universities, medical centers

and laboratories, and conducts research in its own laboratories and clinics.

When laboratory research suggests that a new treatment may be more effective than standard methods, Cancer Clinical Cooperative Groups supported by the Institute conduct studies to evaluate the treatment in cancer patients. These new approaches may include anticancer drugs, radiation therapy and surgery, alone or in various combinations. Many hospitals and medical centers around the country are participating in cooperative group studies.

The current treatment regimens for Wilms' tumor have been validated in a group study under way since 1969, in which members of three cooperative groups and nine unaffiliated hospitals participate. Investigations now in progress will determine whether further reductions in X-ray therapy can be safely made, whether the number of months of chemotherapy can be shortened and, finally, whether there is a role in treatment for an additional anticancer drug recently found active against Wilms' tumor.

In addition to laboratory and clinical research, the National Cancer Institute studies patterns of cancer occurrence in the population. These studies show that although a Wilms' tumor will be seen occasionally in a child in his early teens, 8 out of 10 of these cancers are diagnosed in children under the age of 8. Most are diagnosed in children 3 or 4 years of age.

Children rarely get cancer of any kind. Wilms' tumor is the fifth most common among the types of cancer that do occur each year. About 350 cases are reported annually in the United States.

No causes of Wilms' tumor have been discovered. Research has shown that Wilms' tumor, like all forms of cancer, is not contagious. No one can catch cancer from another person.

There is continuing research on the possible inherited nature of Wilms' tumor. Scientists have not yet reached any definite conclusions on whether or not an inherited tendency exists in the families of most patients. One recent theory is that some cases of Wilms' tumor occur in children who inherited an altered gene from a parent. Although no gene has actually been identified, scientists think that the existence of such a gene might account for cases in which the disease developed at several sites in the same kidney or—more rarely—in the opposite kidney as well, or in a twin or a brother or sister. When patients with these characteristics recover, grow up and have children of their own, the theory predicts that children of this next generation would have an above-average risk of developing Wilms' tumor. Even patients with a single Wilms' tumor might later have children with a slightly increased risk if the theory is correct. Some scientists are working toward development of a simple test to identify people who carry the altered gene—if such a gene is proved to exist.

The recent development of an experimental animal model for Wilms' tumor promises to accelerate research in all aspects of the disease. With the new model, scientists have already isolated a number of *hormones* secreted by Wilms' tumors that might help doctors diagnose the disease and follow the course of treatment. At the

same time, continuing clinical studies offer hope for further gains in treatment.

DEFINITIONS

ABDOMEN	*The part of the body between the chest and pelvis.*
ANIRIDIA	*Absence of the iris of the eye.*
BENIGN	*Not cancer. A benign tumor is a noncancerous growth that does not spread to other parts of the body. Recovery is favorable with treatment.*
CANCER	*A general term for about 100 diseases characterized by abnormal and uncontrolled growth of cells. The resulting mass, or tumor, can invade and destroy surrounding normal tissue. Cancer cells from the tumor can spread through the blood or lymph (the clear fluid that bathes the body's cells) to start new cancers in other parts of the body.*
CATHETER	*A thin, flexible tube used to inject fluid into the body or to drain fluid from parts of the body.*
CHEMO-THERAPY	*Treatment using anticancer drugs.*

CYST	*A sac or vessel containing fluid and foreign matter. Not a tumor.*
DEVELOPMENTAL ANOMALY	*A defect which is the result of imperfect development of the embryo before birth.*
FEMORAL ARTERY	*Major artery in the area of the groin and thigh.*
HEMIHYPERTHOPHY	*Unusual enlargement of half of the body.*
HORMONE	*A substance made and secreted by a gland and carried by the bloodstream to other parts of the body, where it has a specific effect on body functions.*
INTRAVENOUS PYELOGRAPHY	*An X-ray examination of the kidneys, for which a special substance is injected into a vein. Also called* IVP *or excretory urography.*
IVP	*Abbreviation for intravenous pyelography.*
KIDNEY	*One of a pair of organs that regulate the composition of the body's internal fluids and manufacture urine.*
LYMPHATIC SYSTEM	*The circulatory network of vessels carrying lymph, the almost colorless fluid that bathes body cells, and the lymphoid organs, such as the lymph nodes, spleen and thymus, which produce and store infection-fighting cells.*

MALIGNANT	*Cancerous. A growth of cancer cells (see definition of CANCER).*
METASTASES	*Cancer growths that started from cancer cells from another part of the body.*
NEPHRO-TOMOGRAPHY	*The diagnostic procedure using X-rays in which a three-dimensional view of the kidney can be constructed.*
RADIATION THERAPY	*Treatment using high-energy radiation from X-ray machines, cobalt-60, radium or other sources.*
RADICAL NEPHREC-TOMY	*The surgical removal of the kidney, large portions of the surrounding tissue and, often, the neighboring lymph nodes.*
RADIO-ISOTOPE BONE SCAN	*A diagnostic procedure in which a harmless amount of a radioactive chemical is injected into the bloodstream and concentrates in cancer cells. A scanning device passed over the body senses any radioactivity and makes a picture of its location in the body.*
SELECTIVE RENAL ARTERI-OGRAPHY	*A diagnostic method using X-rays and a special chemical to obtain a picture of the kidney blood vessels.*

TUMOR	*An abnormal mass of tissue that may be either benign or cancerous.*
ULTRASOUND	*An experimental diagnostic technique in which "pictures" are made by bouncing sound waves off organs and other structures.*
UREA	*The main waste material in urine.*
VENACAV-OGRAPHY	*A diagnostic method using X-rays to obtain a picture of the major vein that collects blood coming out of the kidney.*
X-RAY	*High-energy radiation used in high doses to treat cancer or in low doses to diagnose the disease.*

Also available:

What You Need To Know About Cancer
What You Need To Know About Cancer of the Bladder
What You Need To Know About Cancer of the Bone
What You Need To Know About Cancers of the Brain and Spinal Cord
What You Need To Know About Cancer of the Breast
What You Need To Know About Cancer of the Colon and Rectum
What You Need To Know About Dysplasia, Very Early Cancer and Invasive Cancer of the Cervix
What You Need To Know About Cancer of the Esophagus
What You Need To Know About Hodgkin's Disease
What You Need To Know About Cancer of the Kidney
What You Need To Know About Cancer of the Larynx
What You Need To Know About Adult Leukemia
What You Need To Know About Childhood Leukemia
What You Need To Know About Cancer of the Lung
What You Need To Know About Non-Hodgkin's Lymphoma
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Office of Cancer Communications
National Cancer Institute
Bethesda, Maryland 20205

National Cancer Institute

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